II.

ELEPHANTIASIS NERVORUM OF THE SCALP: A MANIFESTATION OF VON RECKLINGHAUSEN'S DISEASE.

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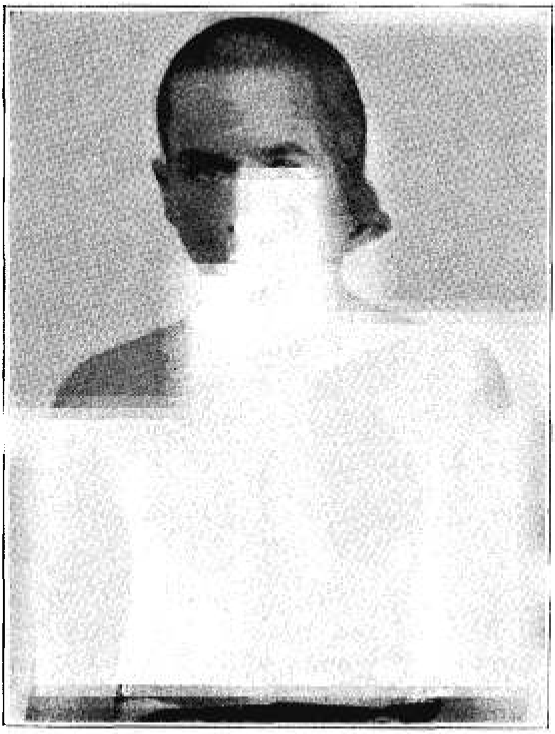
The wisdom of designating maladies, or some of their essential features, by the name of the investigator who first called particular attention to them is contestable. Occasions arise, however, when it is not only a deserved tribute, but a matter of convenience as well. There are few instances in which an individual's name is thus more appropriately linked with a condition of disease than is von Recklinghausen's, for it was he who first demonstrated the histogenetic unity of the morbid changes underlying what had before been considered a number of diverse processes, many of which, owing to their striking and obtrusive nature, had long been known and described under a number of terms. A majority of the terms thus introduced have been descriptive of one or more of the kaleidoscopic features of the malady which now by common consent bears von Recklinghausen's name, but no one term as yet proposed seems to have received general acceptation, generalized neurofibromatosis possibly being as satisfactory as any.

This disease, which, as Feindel has said, is "congénitale toujours, héréditaire souvent, et quelquefois familiale," has for its most constant features certain skin manifestations. These consist chiefly of points or patches of pigmentation, of multiple isolated superficial tumors (molluscum fibrosum), and occasionally of large diffuse growths. In addition to these surface lesions, or indeed in their total absence, there may be multiple tumors distributed along the course of a single nerve or even involving the branches of an entire plexus. These neuromas are often palpable under the skin, but in spite of their size and multiplicity they rarely give rise to pressure phenomena by interrupting the transmission of nervous impulses unless they develop from the nerves during their intraspinal or intracranial course, when compression disturbances of various kinds may arise. Then there occur many so-called symptoms of a secondary order, the evidence of which is less objective: vague sensory disturbances, simple-mindedness, deterioration in mentality, epilepsy, etc.

The patient whose condition we wish to record presented, in addition to many of the commoner features of the disease, a relatively unusual type af diffuse, superficial tumor formation, and the growth, occurring as it did upon the side of the head, was so unsightly that it brought him to the hospital for surgical treatment. Not until we inquired into the matter did we learn that this is by far the more common site of development for these tumors, and the finding has led us to bring together a number of these cases and to present them as a background for our own, the history of which is as follows:

FIG. 1



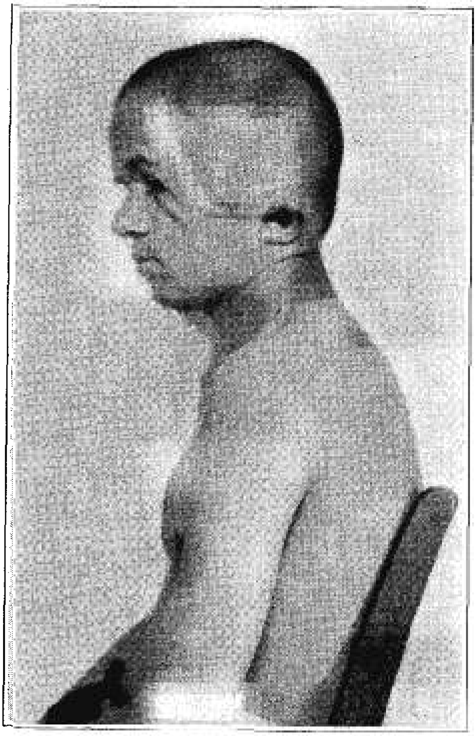
Front view of the patient, showing deformity of the scalp; also areas of pigmentation and other skin lesions over the body.

Thomas P., aged nineteen years, an American, a laborer, was referred to us by Dr. L. P. Hamburger in November, 1905. His complaint was "drooping of the scalp."

Family History. His parents, three sisters, and three brothers are living and well. He is unaware of the existence of any similar trouble in any member of the family, and thinks none of them have any cutaneous blemishes whatsoever.

Personal History. He has always enjoyed good health, and does not recall any of the diseases of childhood. As long as he can remember he has had areas of brown pigmentation and numerous small soft tumors distributed over the body. He does not think that any new ones have appeared during the last few years. It was ascertained from the patient's mother that upon the left temporal region, the seat of the present growth, a large "brown spot" had been present for several years before the skin began to sag. She could not tell whether or not it had been present at birth.

FIG. 2



Side view of the patient, showing displacement of the ear and the outer canthus of the eye.

When seven years of age, while playing in the street, he was run over and was struck by the horse's hoof, on the left side of his head. The skin was not broken, but the scalp was severely bruised.

About a year after this injury the patient thinks the present tumor began to develop. No subjective symptoms accompanied the growth, and as a matter of fact his attention was first drawn to it by his friends, who noticed that his left ear was becoming somewhat lower than the right. This drooping of the ear and sagging of the scalp during the next four years progressed quite rapidly, so much so that the scalp covered part of the left cheek, and the ear stood out at right angles from the head. For the past seven years the patient thinks the growth has been slower. At no time has there been pain or sensory disturbances of any sort associated with this growth. He thinks that hearing in the left ear has become slightly impaired.

Physical Examination. The patient is a listless, undersized young man, who would be taken for a boy of fourteen rather than for a person of his real age. His face is not indicative of a high order of mentality. Although he has attended school, he has never learned to read or write.

On the left side of the head (Figs. 1 and 2), extending from the external margin of the orbit to the occiput, and from a point 2 cm. from the sagittal suture to the left external auditory meatus, there is a soft flabby tumor mass which hangs down from the crown, carrying all of the superficial structures with it. It is very readily movable and slides freely over the underlying bony surface without any evident attachment whatsoever. Palpation of the mass imparts the peculiar sensation which Valentine Mott has likened to a relaxed and emaciated mamma. The scalp sags down over the cheek, so that about one-half of it is covered by hair. The skin of the forehead has been drawn down over the external angular process, carrying the outer canthus with it, partly closing the palpebral cleft, and giving the opening a decidedly downward tilt. Over the crown of the head the tumor is less freely movable than in its more dependent portions. It forms on the side of the face a fold extending from the outer margin of the orbit back to the external auditory meatus. The ear has been crowded so far downward that the tip of the pinna is on a level with the lobe of the opposite ear; it has also been displaced slightly backward, and instead of being parallel to the side of the head the pinna stands out perpendicularly to it. Just above the ear there is a large mass within which several knotted cords can be palpated. The tumor is so freely movable that by picking up a large fold of the redundant tissue over the parietal region and pulling it upward and backward the face can be restored almost to its normal outline. The scalp over the tumor is greatly thickened, its surface roughly papillated, and in several areas covered by patches of brownish pigmentation. The hair over the tumor is thinned out and in many follicles only short broken stumps of coarse hair are to be seen. Many of these hairs are without pigmentation. The patient's beard has not as yet developed; merely a few downy hairs are to be seen.

There seems to be some slight atrophy of the temporal muscle under the tumor with relaxation of its fascia, for the index finger can be readily introduced for some distance behind the zygoma. Posterior to and somewhat below the level of the ear are two smaller tumors having the same brownish color and the same consistency as the larger tumor mass. They measure about 3 x 4 and 2 x 3 cm. respectively. During the examination slight spasmodic twitchings of the facial muscles were seen to occur.

Over the thorax and abdomen the skin shows a variety of lesions. Scattered quite profusely over the trunk, and the extremities as well, are small areas, pin-head in size, of brownish pigmentation. There are also larger patches of pigmentation of irregular outline (Fig. 1), the larger ones occurring upon the chest, the smaller ones scattered over the body, even upon the hands and fingers. There are no patches upon the palms. There is no evidence of segmental distribution in the topography of these patches. Especially over his chest and flanks are scattered numerous slightly elevated bluish discolorations, resembling minute bruises. Over the chest and back also are numerous so-called spider angiomas. Many small moles, both sessile and pedunculated, are scattered over the body. His back in particular is peppered with these small growths. Finally, on the chest, one arm, and one leg are a few characteristic soft fibromas. These latter tumors are all of the same general character, of a brownish color, slightly raised above the surface, soft and compressible, giving the characteristic seedless-raisin feeling when examined. They measure on an average about 1 cm. in diameter.

FIG. 3

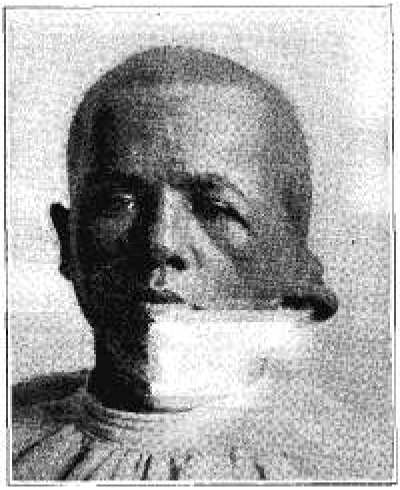
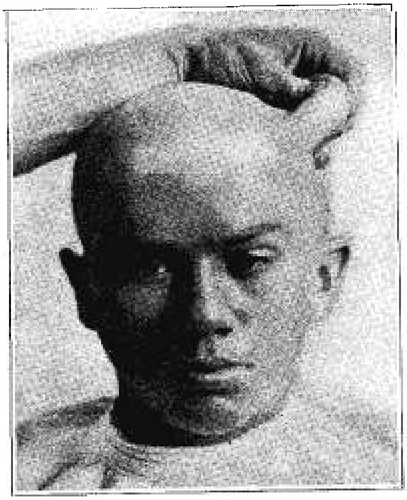


FIG. 4



The condition after preparation for operation, to show drooping of the scalp, with displacement of the ear and the outer canthus, and replacement of these on gathering up a fold consisting of the main part of the tumor growth.

A careful examination of the palpable nerves of the extremities discloses no irregularities in their structure. In only one place are any definite neuromas of nerve trunks found. These are associated with the large tumor, and will be described later.

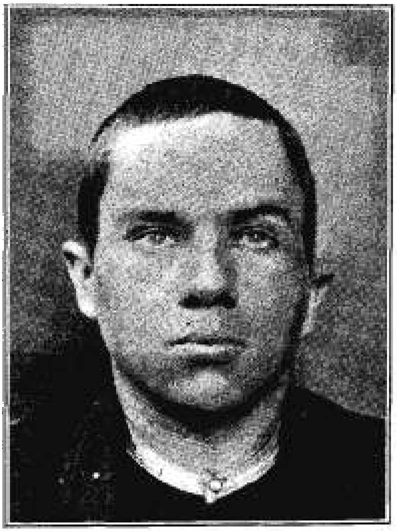
Operation under ether November 9, 1905. The accompanying photographs (Figs. 3 and 4) of the patients head were taken by Dr. Gilman, after he had been shaved in preparation for the operation.

After cleaning up the scalp, and while palpating the temporal vessels to determine whether a tourniquet could be so applied as to shut off the extracranial blood supply, the operator found a nodule the size of a bean lying just above the zygoma in front of the ear, evidently upon the auriculotemporal branch of the fifth nerve. Above this nodule could be felt a chain of others, running out into the tumor mass. Consequently, before attacking the tumor proper an incision was made over this region and the primary nodule, while the secondary ones lying above it were dissected out, the temporal artery being ligated at the same time. The tissues were found to be soft, almost gelatinous, and exceedingly vascular, so that some difficulty was experienced in checking hemorrhage even in this small wound. It was closed by deep mass sutures.

The flap of scalp which was to be removed was then pulled up so as to restore slightly more than the natural position of the ear and the horizontal position of the palpebral cleft. A tourniquet was then applied, holding the scalp in this position. The proposed area of tissue for removal was then marked out with the scalpel by a superficial scratch through the epidermis. The incision extended from a point near the external angular process nearly to the occipital protuberance, and included an area of skin on its surface about 15 cm. in cross diameter. It is well that we took this precaution, because from the first deep incision the bleeding was so profuse that had this preliminary step not been taken it is probable that just the right amount of tissue to ensure a successful plastic result would not have been removed, owing to the confusion which the abundant hemorrhage occasioned. As rapidly as possible the incision was carried down through the scalp to the cranial aponeurosis, and as the tissue was dissected away from before backward, the edges of the wound were brought together en masse by a continuous Pagenstecker suture. No vessels were ligated, for we found that the clamps pulled out of the soft tissue by their own weight as rapidly as they were put in. It was only by this taut closure of the wound that we succeeded in controlling the bleeding. The edges of the incision were then more leisurely approximated by interrupted fine silk sutures. No atrophy or change in the bones or the pericranial tissues was disclosed during the operation.

Though the patient lost an amount of blood during the operation sufficient to leave him somewhat anemic for a time, he made a rapid and satisfactory recovery. At one point the sutures in the scalp had failed to check the bleeding completely, so that a small hematoma formed underneath the edge of the incision. When the sutures were removed some days later this clot broke through and left a point which healed slowly by granulation. The practical restoration of the eye and ear to their normal position is shown in the accompanying photograph (Fig. 5). It is quite possible that there may be a recurrence of the growth, particularly as the two isolated masses resembling the original tumor and lying over the mastoid process were not removed with the spindle-shaped piece of tissue taken away at the operation. For a few weeks after the operation the angle of the eye and the ear were drawn up into a position somewhat higher than on the opposite side, but this overcorrection of the pre-existing deformity has gradually disappeared.

FIG. 5



Postoperative condition, to show restoration of the eye and the ear to practically the normal position.

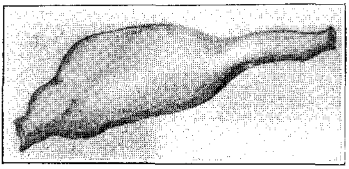
Pathological Findings. The tissues for examination were a neurofibroma removed by a separate incision from just above the left ear, and a diamond-shaped piece of scalp from the left parietal region.

The neurofibroma measured 1.8 x 0.8 x 0.6 cm.; it was soft and elastic, and of a dull-gray color. The surrounding tissue readily stripped from its capsule, leaving a smooth and glistening surface. Toward one end there was a small sulcus which indefinitely divided the tumor into two lobes. The nerve trunk proximal to the neurofibroma was uniformly thickened as far as it had been removed; the nerve distal to the tumor was of smaller diameter, and it emerged from the main lobe (Fig. 6). On fresh section the tumor was translucent, smooth, rather œdematous, and of a yellowish-gray color. Running through its centre was to be seen a more opaque, whitish band, striated transversely, and at one point sending off a branch into the other of the two lobes. The capsule was everywhere well defined.

Microscopically, the neurofibroma is found to be made up for the most part of a loose connective tissue. It consists of interlacing strands of fibers forming a very fine network, the meshes of which appear almost like vascular spaces. This loose tissue varies considerably in its cellular contents; in some areas it is almost entirely fibrous, and in others very cellular. Through the centre of the tumor there passes a dense band of connective tissue, in the wavy outlines of which the nerve fibres can be demonstrated.

The central strand is made up principally of spindle-shaped cells with large vesicular nuclei. The looser tissues consist of finer drawn-out cells with solid, dark-staining nuclei. Occasionally very large vesicular nuclei are to be seen. There are very few cells of the wandering-cell type, although an occasional group of young fibroblasts resembles them very closely. Though there are but few vessels in the tumor itself, the vascular supply of the capsule is abundant, and several tags of adherent tissue prove to be of the same type as the tissue which makes up the greater part of the thickened scalp.

FIG. 6

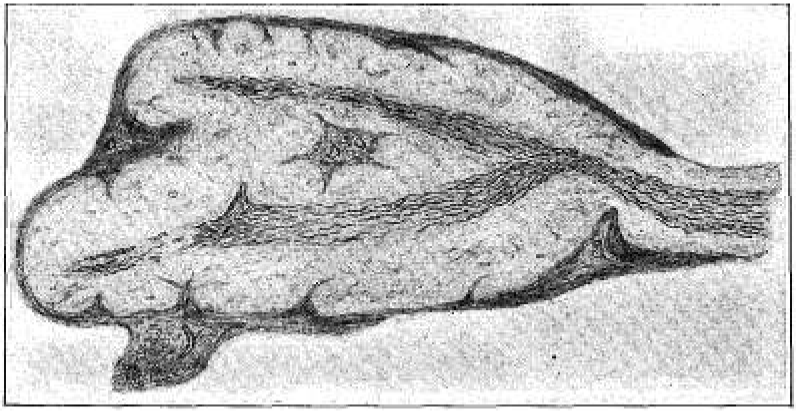


Sketch of the neurofibroma from the auriculotemporal branch of the fifth nerve. (Actual size.)

With Mallory's connective-tissue stain the central cord stands out very prominently, and among the blue connective-tissue fibers a few yellow-stained nerve fibers can be made out. Sections stained with Weigert's elastic-tissue stain gave negative results. The specific stains used to demonstrate the nerve fibers were Weigert's myelin-sheath stain and Robertson's modification of Heller's stain. The latter stain, on account of its more definite color contrast, was used in the great majority of the preparations. A series of sections taking in the entire diameter of the neurofibroma was stained. The series shows that there are two main strands of nerve fibers running through the tumor mass, about as they are indicated in Fig. 7, which does not represent any one section, but a composite of one near the beginning and one near the end of the series, in order to bring out in a clearer manner the course of the fibers. As can be seen in the sketch, the greatest part of the tumor is due to a proliferation of the perineural connective tissue, but judging from the distance which separates the individual nerve fibers there has been considerable proliferation of the endoneurium as well. The amount of endoneural thickening varies considerably in different parts of the tumor. The fibers of the entering nerve are quite widely separated, while the nerve leaving the tumor shows some perineural thickening, but the nerve fibers themselves form a compact bundle. The point at which the second band of fibers leaves the tumor corresponds to the small lobe that was made out in the gross. The nerve evidently was cut off short. The individual nerve fibers, both large and small, appear perfectly normal.

Scalp. In its shrunken condition after hardening the portion of scalp removed measured 15.2 x 7.5 cm. Its thickness varied from 0.5 cm. at the periphery to 1.2 cm. in the central areas. The epithelial surface was irregular, due to marked papillary formation about the hair follicles. This was especially apparent in those areas where brownish pigmentation was most abundant. The cranial surface was rather ragged in appearance, and the tissue was quite uniformly soft, œdematous, and of a yellowish-red color. It had a peculiarly flabby feel to it. At the posterior inferior corner of the spindle-shaped piece of tissue there were found several nerves which showed nodular thickenings. One of these thickenings measured 1.4 cm. in length by 0.2 cm. in diameter. These were the only neurofibromas seen on inspection, and are indicated at *a*, in Fig. 8.

FIG. 7



Longitudinal section of the neurofibroma pictured in Fig. 6. (Robertson's modificatian of Heller's stain.)

The thickening of the scalp was due almost entirely to œdematous connective tissue, which after hardening in formalin was quite tough and could be readily pulled off in sheets, and sometimes also in small cords that resembled nerve fibers. In the areas of greatest thickening the nerves and neurofibromas were no more abundant than elsewhere.

On cross-section of the scalp the tissue presented a uniform fibrous appearance. The hair follicles were enlarged and extended in some areas three-fourths of the distance through the scalp. About them there was a small ring of fibrous tissue. The bloodvessels that were cut in different sections were not loose in the subcutaneous tissue, and a definite wall could not be dissected out, but were firmly embedded in the fibrous tissue. One vein was cut longitudinally for a distance of about 2.5 cm. It did not collapse, but remained open its entire length, looking like a small trough. In attempting to free it from the surrounding tissue it was torn, but could not be dissected free.

FIG. 8

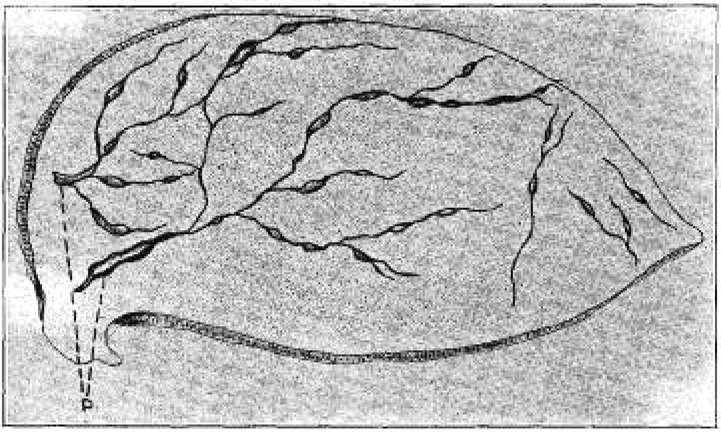


Diagram of the neurofibromas, dissected out in the removed portion of the scalp.

On carefully dissecting out some of the nerve twigs found on the under surface of the scalp numerous small neurofibromas were found. These tumors were all of about millet-seed size, scattered irregularly along the different nerves, as shown in Fig. 8. They were most abundant just beneath the corium, lying between the hair follicles (Fig. 9). The nerve fibers became so fine in this layer that after the cluster of neurofibromas was dissected out it was almost impossible to follow them further. In one instance after a nerve had divided into two branches a neurofibroma on one of its branches, about 0.5 cm. beyond the division, enclosed the other branch as well. The smaller tumors had the same cross-striation that was seen in the central cord of the neuroma described above at some length. In every instance the neurofibromas, owing to their definite encapsulation, could be readily shelled out from the surrounding fibrous tissue.

On microscopic examination of the scalp the most marked change to be noted is the great increase in cellular connective tissue in the subcutaneous layer. This tissue shows no definite arrangement, but consists of dense, irregular masses, chiefly in the lower layers of the tela subcutanea. In many areas also the normal fibrous corium is replaced by this same cellular connective tissue, which in many spots may extend even as far as the epithelium itself. The tissue consists of fibroblastic cells with relatively large vesicular nuclei, that run in bands, layers, or form an irregular network. Numerous "mast-zellen" are scattered through the connective tissue. The subcutaneous tissue contains a considerable amount of fat, which also has been invaded by the sclerosing process, for the alveoli are separated by thick strands of fibrous tissue.

Sections stained with Weigert's elastic-tissue stain demonstrate very clearly that the portion of the scalp below the corium is relatively free from elastic fibers. Throughout the pars reticularis and papillaris of the corium itself, however, a fine tracery of elastic fibers can be demonstrated. At numerous points small strands of tissue containing elastic fibers extend from the corium for short distances into the subcutaneous tissues. In the tela subcutanea proper the elastic fibers are very poorly developed; only here and there are seen a few small scattered fibers. In the lowest portion an occasional fiber is seen.

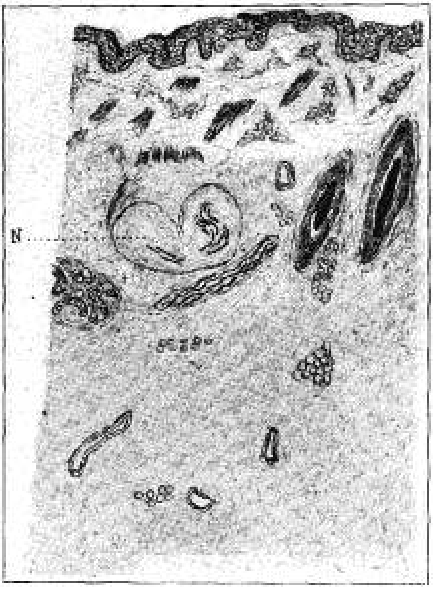
The epithelium is normal in appearance, except for its unevenness, which was noted in the gross. Underlying the epithelium the cells contain some yellow pigment, as do also the wandering cells in the spaces of Lange. The corium, except for the invasion of fibrous tissue from below, also appears normal.

The hair follicles are larger than normal, dipping deep down into the subcutaneous tissue, and the connective tissue about them is increased in amount. The sebaceous glands are also proportionately enlarged. The sweat glands are not increased in size or number, nor are there to be seen any finger-like processes growing into the subcutaneons tissues, such as von Recklinghausen described. The capsules of the glands are slightly thickened.

The lower layer of the subcutaneous tissue is quite vascular. The walls of the vessels do not stand out distinctly, as is the rule in connective tissue, but they are firmly embedded and seem fused with the cellular connective tissue about them. This is especially well seen in some of the larger veins, in which the wall proper cannot be distinguished from the concentrically arranged connective tissue, which at certain points seems almost to make up the entire wall. The elastic tissue in the vessel walls seems normal.

The neurofibromas were very infrequent in the different sections taken from the scalp, though numerous nerves were seen, showing slight increase in the size of their perineural sheaths. One of these neuromas is shown in Fig. 9. The nerves, as well as the tumors upon them, are definitely separated from the surrounding fibrous tissue, sometimes even by a small interval. Thus, the interlacing fibrous tissue cells of the nerve sheath are quite distinct from the surrounding tissue which possesses the same histological character. The nerve fibers in the small subcutaneous tumors are few and run irregularly through them, so that very probably there has been a proliferation of the endoneurium as well as of the perineurium, a condition similar to that found in the larger neuroma first described.

FIG. 9



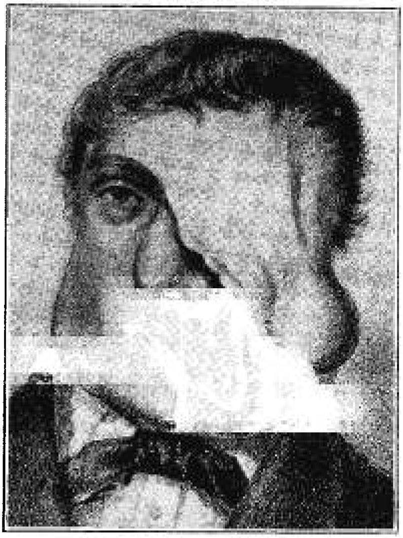
Miliary neurofibroma in a section of the scalp. (Robertson's modification of Heller's stain.)

Just why the trigeminal field and the temporal region in particular should be the seat of predilection for these growths is not apparent. Bruns, in his review of these cases, found that more than a third of the so-called "Rankenneuromata" originated in this territory. The tumors are painless, and troublesome only because of their unsightliness and the great size which they may attain. Hence, it is natural to suppose that they will more often lead their hosts to seek surgical relief when they occur upon an exposed part of the body than when they are hidden from sight. But even this does not suffice to account for the great predominance of the temporal cases. They are, furthermore, much more common in men, as the subjoined list indicates.

Only one of the several patients afflicted with von Recklinghausen's disease who have been admitted to the various services at the Johns Hopkins Hospital has possessed an elephantiatic growth of this nature. This was a woman with all the hall-marks of von Recklinghausen's disease, in whom an enormous tumor mass hung from the right side in much the same position and of about the same proportions as in the case depicted by Virchow as a frontispiece to *Die Krankhaften Geschwülste,* vol. i. This, after the temporal region, is perhaps the second most frequent seat of origin for these tumors.

Although these large isolated growths are almost invariably accompanied by other manifestations of von Recklinghausen's disease, the tumors themselves have come to be designated by a most variable terminology. In his classical monograph, von Recklinghausen himself adheres to the term elephantiasis mollis, used by Virchow. Mott designated the growths pachydermatocele. Verneuil introduced the terms plexiform neuroma and neuroma cylindricum plexiforme. Among other designations are elephantiasis molluscum (Nélaton); rankenneurom and neuroma circoideum (Bruns); dermatolyse (Marie); elephantiasis neuromatodes (Bruns); lappen-elephantiasis (Esmarch); cylindrisches fibrom (Marchand); fibroneuroma racemosum (Rizzoli); tumeur royale (Boudet), etc.

FIG. 10



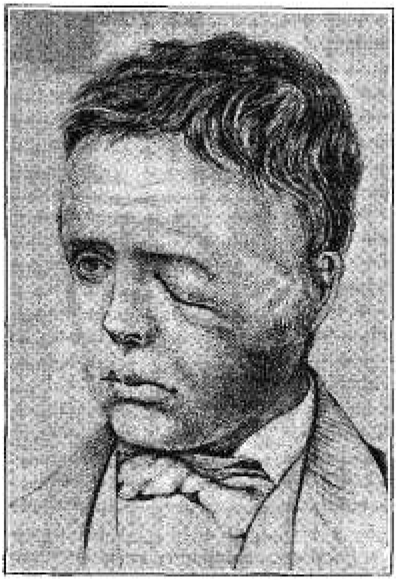
Valentine Mott's case of "pachydermatocele."

In his monograph von Recklinghausen cites one or two cases which appeared in the literature before 1854, but possibly it may be well to begin our report with the cases which were recorded by Valentine Mott.

1854. Valentine Mott. "Tumors of Skin Denominated Pachydermatocele," *Med. Chir. Trans.*, London, 1854, vol. xxxvii, p. 155.

Case III of his series. A boy, aged fourteen years, consulted him because of a "hideous deformity" on the right side of his head and face. The tumor formed three layers, extending from the crown of the head to below the jaw. One fold involved the upper eyelid, and when raised revealed the globe at the end of a canal four inches in depth (Fig. 10). The tumor was twice operated upon, but the growth recurred after each operation.

FIG. 11



Billroth's Case of "plexiformes neurofi brom."

Case IV of his series. A boy, aged twelve years, presented a swelling on the right side of the face which began in early infancy and was probably congenital. Its description corresponds exactly to the case cited above. The operation was successful and there was no return of the growth during the following six years.

It is to be noted that Mott limited the description in these cases to the tumor lesion alone, and he makes no reference to the presence of other skin manifestations which we now include under the term von Recklinghausen's disease.

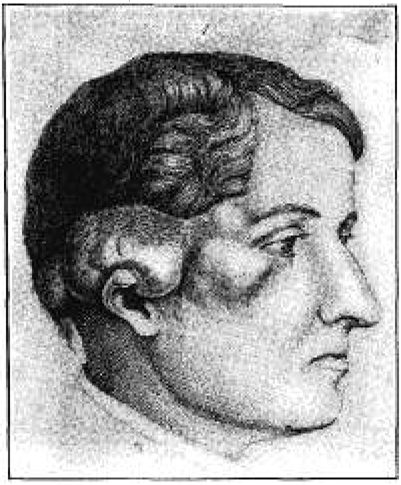
1863. Billroth. "Ueber die Entstehung der Fibroide," *Archiv f. klin. Chir.*, 1863, Band iv, p. 547.

A boy, aged six years, presented a tumor consisting of small, hard strands and masses overlying the right upper eyelid and temporal region. On microscopic study of the tumor after its removal it was found that there was a central strand, evidently a nerve, which on entering the growth became either fibrous or changed into a soft mass of fat. Its structure could not be clearly made out.

1869. Billroth. "Plexiformes Neurofibrom des oberen Augenlides und der Schläfengegend," *Archiv f. klin. Chir.*, 1869, vol. xi, p. 232.

A boy, aged eighteen years, strong and healthy, of moderate mental development, had had since birth a tumor of small size over the temporal region. It had suddenly taken on a rapid growth unaccompanied by any symptoms of discomfort. It had become as large as a fist, and involved the temporal region and upper eyelid. It was soft and flabby, and extended deep into the orbit. A defect was found in the parietal bone about the size of a pea. The tumor, which was removed, was seen to consist of a mass of plexiform, grayish-red, smooth, round cords. During the following thirteen months there was no recurrence (Fig. 11).

FIG. 12



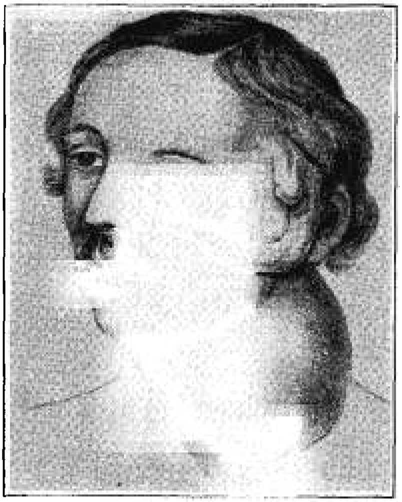
Bruns' Case I, "Ranken-neurom."

1870. Bruns. "Das Ranken-Neurom," *Virchow's Archiv*, 1870, vol. l, p. 80.

Case I, a man, aged twenty-eight years, had had since birth a painless tumor in the right temporal region. There was no history of a similar affliction in the patient's family. There were no intellectual disturbances; his general health was good. The tumor was situated just above the right ear, which was so pushed down that the pinna stood out horizontally (Fig. 12). The tumor possessed a wormy feel, and was of rather soft consistency. It was removed, the patient dying from infection some days later. Microscopic examination showed new formation of nerve fibers by metaplasia of connective tissue.

Case II, a patient, aged thirty-three years, whose brother, Case III, had a similar tumor. The mother had numerous cutaneous wart-like excrescences over her body. Since birth the patient had had a flat tumor involving the left temporal region and left upper eyelid. At the time of puberty the growth developed and the eye had been lost owing to a suppurative infection. There were several painful spots over the tumor. On palpation numerous coils of a firmer consistency could be made out in the soft stroma of the growth.

FIG. 13



Bruns' case II, "Ranken-neurom."

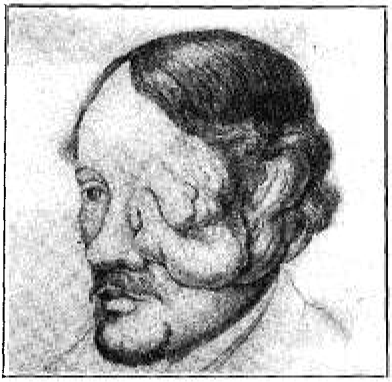
As seen in the illustration (Fig. 13) there was also a large cervical tumor which was removed at operation. The patient died on the tenth day from hemorrhage due to ulceration of the carotid artery.

Case III, a man, aged forty-three years, a brother of Case II. This tumor likewise was congenital and resembled very closely the one described above (Fig. 14). The eyeball in this case also had been lost at the time of puberty. There were no local areas of tenderness. The tumor was soft and movable, and on deep palpation numerous so-called cords could be found running through it.

1870. Laroyenne. Christot, *Gaz. hebdom.*, 1870. Ref. Cartaz, *Archiv. gén. de Méd.*

A child, aged five years, healthy in appearance, presented a painless tumor over the right cheek, which had enlarged rapidly shortly before he came under observation. The tumor was soft and had numerous small palpable thickenings scattered through it. An incomplete operation was followed by immediate recurrence. The microscopic characters were those of a plexiform neuroma, showing at many points sarcomatous degeneration.

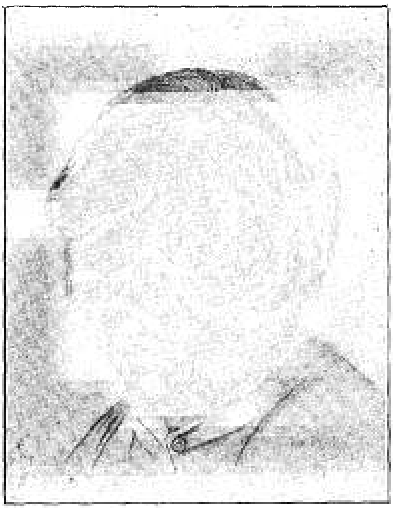
FIG. 14



Bruns' Case III, Ranken-neurom. Brother of Case II, Fig. 13.

1870. Rizzoli. *Lo Sperimentale,* November, 1870. Ref. Bruns, *Beiträge zur klin. Chir.,* 1891, vol. viii, p. 1.

FIG. 15



Billroth's case of bilateral involvement of frontotemporal region.

Two cases are reported by Rizzoli, one of them a child, aged six years, in whom a tumor growing from the upper eyelid and skin over the orbital arch had been present since early life. In the other patient, a man, aged twenty years, there had been present since early life a ranken-neurom which had developed in the temporal region and upper eyelid.

1870. Billroth. *Chir. Klinik,* Wien, 1869–1870, 54.

A man, aged thirty-three years, had had since early life a small swelling over the left eyebrow. Later on a similar swelling appeared over the right eyebrow. These tumors had slowly increased in size, forming sac-like folds which hung down over the eyes and cheeks (Fig. 15). After twenty operations the greater part of these tumor masses was removed. The patient had also a few superficial pea-size tumors over his chest.

1873. Fritsche. "Two Unusual Cases of Elephantiasis Arabum," *Clin. Soc. Trans.,* London, 1873, vol. vi, p. 160.

A patient, a beggar, aged twenty-six years, had an elephantiatic transformation of the right half of his face; "the whole side of his face resembled an empty hanging bag." The tumor had been present since childhood. The opening of the right eye was on a level with the nostril. The eyeball was atrophied and hung by a long pedicle, the optic nerve being very near the opening of the eyelids. The helix of the right ear was enlarged and hung loosely down. The patient was said to be otherwise normal.

1875. Stokes. "Pachydermatocele or Fibromolluscum," *Dublin Journal of Med. Sci.,* 1875, vol. lix, p. 69.

A man, aged thirty-two years, had had a slowly growing tumor situated in the temporal region. It had appeared early in life and had grown slowly. His general health was good. The tumor was very movable; its surface was irregular and nodular. It was not discolored, but was thickly covered with hair. An operation was successfully performed, though there is a note of great difficulty from a tremendous hemorrhage.

1876. Billroth. "Geschwülste," *Chir. Klinik,* Wien, 1872–1876.

A girl, aged fifteen years. There had been noticed since childhood a fulness of the right temporal region. In this situation a tumor developed which involved the temporal region and upper eyelid and upper part of the cheek. In the tumor mass numerous hard, irregular cords could be palpated. The tumor was tender. In two operations it was completely removed and there was no recurrence.

1876. Cartaz. "Etude sur le Neurome Plexiforme," *Archiv. gén. de Méd.,* 1876, vol. cxxxviii, p. 176.

A boy, aged eleven years, with a negative family history, had had since birth a tumor, the size of a dollar, elevated about 1 cm. above the scalp. It was situated over the left ear. It had developed slowly until within a few months, when it had doubled in size. It was painless. It hung down over the neck, and in the soft mass could be palpated cords and chains of harder tissue. The operation for its removal, though successful, proved a very bloody one.

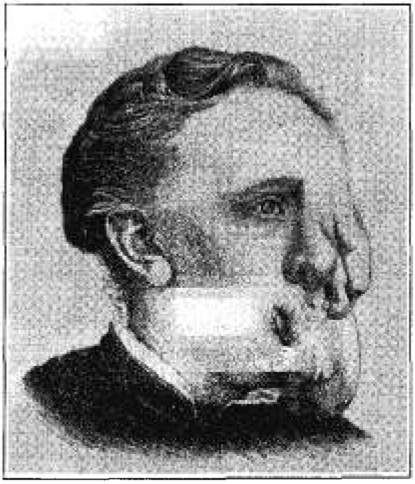
1877. R. Marchand. "Das Plexiforme-Neurom (Cylindrische

Fibrom der Nervenscheiden)," *Virchow's Archiv,* 1877, vol. lxx, p. 36.

A boy, aged twelve years, in whom a tumor was first noticed when six months old. The growth was very slow. There was no history of a similar condition in the family. The upper lid of the left eye and adjacent part of the temporal region were involved. The upper lid hung down like a sac over the left eye, but on raising the tumor mass the eye was found to be normal in all respects. On deep palpation numerous dense cords could be made out. Deep pressure elicited some pain. The patient was successfully operated upon and the tumor is said not to have recurred.

Marchand reports the case of a boy aged eighteen years. There had been present since his fourth year a large tumor situated just above his right ear. It had slowly enlarged, and at the time of operation consisted of a soft, doughy, painful swelling over the right temporal and occipital regions. In the depths were felt numerous strands of firm tissue on which small enlargements were palpable. The tumor was partially removed by operation.

FIG. 16



Schüller's case, involving the territory of the superior maxillary as well as the frontotemporal region.

1878. Schüller. *Deutsche Zeitschrift f. Chir.,* vol. ix, p. 261.

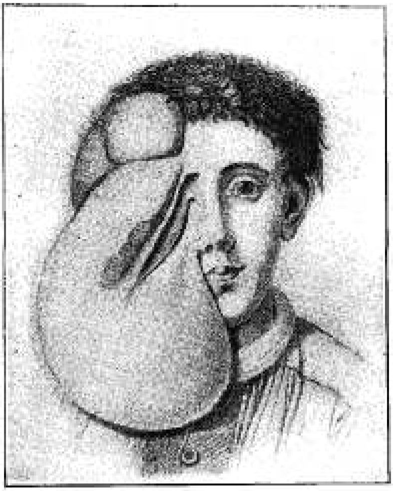
A woman, aged eighteen years, presented a tumor involving the left side of the face. It had developed slowly until puberty, after which time its growth was more rapid. The mass presented the usual characteristics of position and consistency (Fig. 16). It was removed, with a fair cosmetic result. The eyeball had been destroyed by suppuration some time before the operation.

Schüller reports a second case in a woman aged twenty-four years. A tumor on the right side of the face had been present since infancy, enlarging slowly until the time of puberty, when it had rapidly increased in size. In the deeper portions of the tumor several cords and knots could be palpated. It was successfully removed at operation.

1880. Schultze. "Ein Fall von sehr grosser Fibroma Molluscum an Kopf und Gesicht," *Deutsche Zeitschr. f. Chir.*, 1880, vol. xiii, p. 374.

A young man, aged twenty years, had a tumor the size of his head hanging from the temporal region and cheek. It measured 31 x 22 x 12 cm. in its several diameters. There was no family history of a similar condition. The growth was first noticed when the patient was four years of age. It was then about the size of a pea. Within a year it grew considerably and was removed. It returned almost immediately and rapidly enlarged to its present size (Fig. 17). A radical operation was done in three stages, with a fair cosmetic result.

Fig. 17



Schultze's case, designated a large "fibroma molluscum."

This case was studied histologically by von Recklinghausen, who was unable to demonstrate any nerve fibers in the tissue, but he considered this to be due to the enormous size of the growth, which was made up of fibrous tissue at the expense presumably of a few nerve fibers which were not demonstrable. He nevertheless regarded the case as one of "elephantiasis mollis."

1882. Labbé. (Case IV of Bruns' series.) *Beiträge zur klin. Chir.*, 1891, vol. viii, p. 1.

A woman, aged twenty years, had a tumor involving the forehead and upper eyelid, which had been present since she was a year old. The growth hung loosely down in a large fold.

1880. Schuster. (Case XVI of Bruns' series (*loc. cit.*). A boy, aged fourteen years, had an elephantiatic enlargement which had been present since his fourth year. It involved the nose and cheek. The body was covered with numerous areas of pigmentation and fibroma molluscum.

1882. Socin. "Angeborne Elephantiasis der Rechten Gesichtshälfte," *Jahresbericht der Chir.*, Abth. d. Spitals zu Berne, 1882, p. 29.

A woman, aged twenty-seven years, who had always been healthy presented a tumor which had been slowly growing and which covered the right half of the forehead and temporal region, surrounding the orbit like a crescent, and hanging over the region of the upper jaw. It was pigmented, and within the mass could be felt the usual knotted cords. The upper lid was greatly enlarged and hung down over the eye in a large fold. In spite of serious hemorrhage the tumor was successfully removed in two stages.

1883. Volkman. "Beiträge zur Kenntniss vom Plexiforme-Neurom," Dissertation, Magdeburg, 1883.

A boy, aged ten years, had a congenital tumor hanging from the scalp, which reached as low down as the clavicle. The skin over the tumor was pigmented, hypertrophic, and was thickly covered with hair.

1890. Rapok. "Beiträge z. Statistik der Geschwülste," *Deutsche Zeitschr. f. Chir.*, vol, l. S. 538.

A girl, aged seventeen years, showed a tumor which developed in her upper eyelid during childhood. For a year and a half before she came under observation the growth had been much more rapid. Her father also had "neurofibroma plexiforma."

1891. Andry and La Croix. *Lyon Médical,* 1891, tome lxvii, No. 21, p. 109; No. 22, p. 145.

A boy, aged eight years, giving no history of a similar condition in the family, had a tumor which was first noticed when he was two years of age. He was of normal mentality. The growth had been gradual and at the age of eight the tumor, involving the left temporal region and the left upper eyelid, had reached a large size. It presented the usual characteristics. It was successfully removed at operation.

1891. Bruns. "Ueber das Rankenneurom," *Beiträge zur klin. Chir.*, vol. viii, p. 1.

Case VII of his series. A female, aged twenty years, had a tumor of congenital origin, involving the upper eyelid and orbit. The upper lid was greatly enlarged and elephantiatic. There was also a defect in the roof of the orbit.

Case XI of his series. A patient of Dessauer's. A man, aged forty-two years, had a tumor of the temporofrontal region and involving the upper eyelid. There was also a large growth of similar character hanging from the loin. The tumors consisted of several soft flabby folds of skin. Besides these tumors he had numerous nævi and fibroma molluscum.

Case XIV of his series. A man, aged thirty-eight years, had a congenital tumor involving the frontal region and upper eyelid. Hanging from the forehead, the tumor was draped down over the eye in several soft, flabby folds. There were numerous fibromata molluscum scattered over the body.

Fig. 18

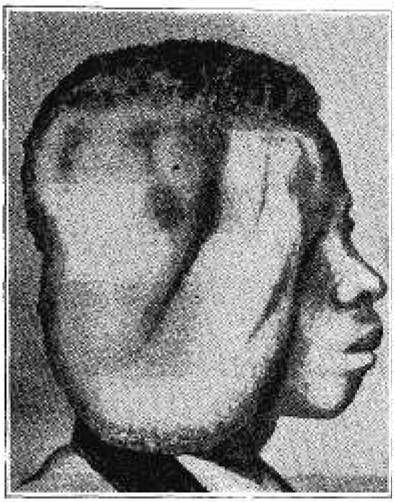
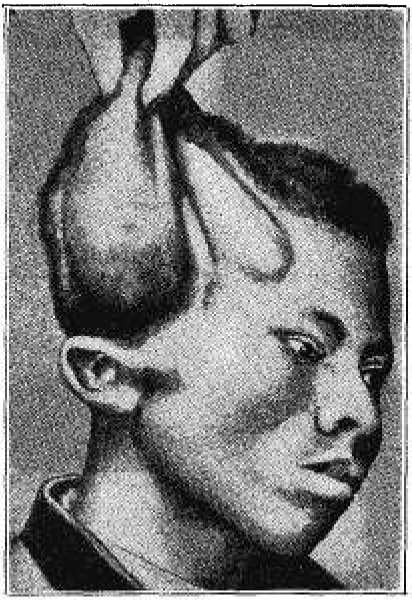


Fig. 19



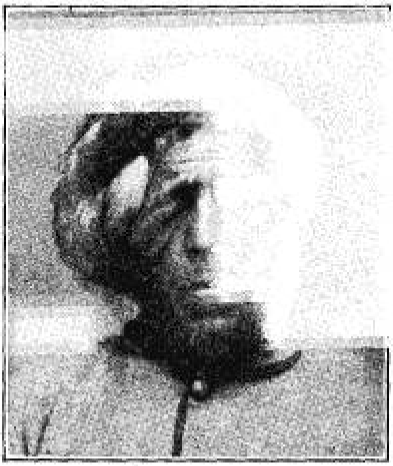
Magalhaes and Manson's case of "congenital elephantiasis."

1893. Magalhaes and Manson. "A Case of Congenital Elephantiasis of the Scalp," Amer. Jour. Med. Sci., 1893, vol. cv, p. 120.

A man, aged thirty-two years, since childhood had a growth which originated from the right side of the scalp and had slowly and progressively increased in size. The "hood-like" mass was soft and flabby. The right half of the cranium corresponding to the tumor was markedly deformed (Figs. 18 and 19).

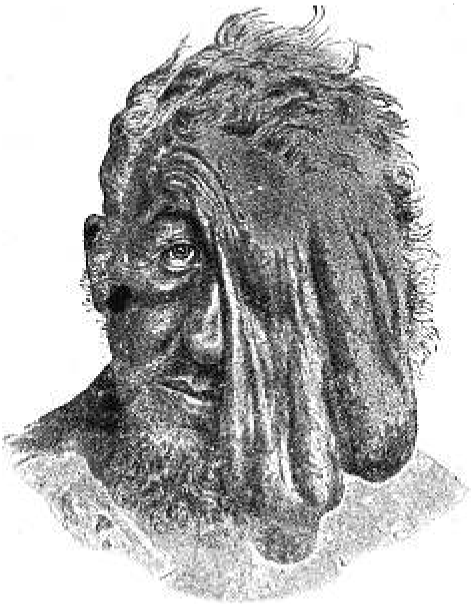
1896. Dennis. We are indebted to Dr. Frederic S. Dennis for this photograph (Fig. 20) of a patient whose tumor and other skin lesions are depicted in a more advanced state in vol. iv. of his *System of Surgery*. The condition is associated with the other manifestations of von Recklinghausen's disease, and the growth was of long standing.

FIG. 20



Dennis' case.

FIG. 21



Lanz's case of "fibroma molluscum."

1904. Lanz. *Handbuch d. Chir.* Bergmann, Bruns, and Mikulicz, second edition, vol. i, p. 31.

A man, aged twenty years, had a large growth (Fig. 21) which made him look as though he was wearing a pendulous cap. The mobility of the mass was very marked. He had fibroma molluscum and areas of pigmentation scattered over his entire body. The tumor was successfully removed.

1905. Wynn. "Plexiform Neurofibroma," *Jour. Amer. Med. Assoc.*, 1905, vol. xlvi, p. 500.

A young man, aged nineteen years, giving no history of similar affliction in the family, had an elephantiatic tumor of the forehead, which was removed when he was eleven, but which gradually returned. Beginning about the fourth year, there developed a dense, diffuse enlargement over the parietotemporal region, and somewhat later in the orbits and cheek. The skin hung in folds from the orbits down over the lower jaw. Irregular, worm-like masses could be palpated deep in the tumor. Microscopically, no nerve fibers were demonstrated.

ORIGINAL ARTICLES.

GRAPHIC METHODS IN THE STUDY OF CARDIAC DISEASES.[1]

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(From the Medical Service of the Johns Hopkins Hospital.)

The first observations upon pulsation of the veins was reported to the French Academy by Homberg in 1704,[1] in a case of heart failure; Zuliani, in 1805, and Benson, in 1836, noted similar pulsation in cases in which autopsy showed tricuspid insufficiency; and Kreysig, in 1814, observed the pulsating liver in such cases. The experimental study of the pulsation in the veins was first undertaken by Barry in England, in 1826, and by Wedemeyer in Germany, in 1828;[2] but although Weyrich, in 1853,[3] and Bamberger[4] and Friedreich,[5] a decade later, made further contributions to this subject, little practical importance was attached to it. The scientific investigation of pulsations and pressure changes in the circulatory system really dates from the publication of Marey's *Physiologie médicale de la circulation du sang* in 1863, and the careful and complete experimental and clinical studies emanating from his laboratory have furnished the basis for most of the work done before 1895. Since then the work of Engelmann[6] on the myogenic origin of the heart-beat, and its clinical application by Hering[7] and Wenckebach,[8] have given a new stimulus to the study of the diseased

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[1] Read before the Johns Hopkins Medical Society, Baltimore, February 19, 1906.